

Congenital double chambered left ventricle treated by exclusion of accessory chamber

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SUMMARY A 9 month old boy with a double chambered left ventricle presented with congestive cardiac failure refractory to medical treatment. He was successfully treated by surgical exclusion of the accessory chamber.

The double chambered left ventricle is a rare condition and to our knowledge only six cases have been previously reported¹⁻⁴. We present the first case of this condition successfully treated by exclusion of the accessory chamber.

Case report

This child was born at 38 weeks gestation after a normal pregnancy. His birthweight was 3.2 kg.

He remained well until the age of 3 months when he started to experience feeding difficulties, and at the age of 4 months he was admitted to the Royal Belfast Hospital for Sick Children in congestive cardiac failure. This was treated with digoxin 0.03 mg bd, frusemide 10 mg bd, and aldactone 6.25 mg bd. Cardiac catheterisation at this time showed a double chambered left ventricle (Fig 1). There was free communication between the two chambers, both of which contracted poorly. The pressures in the two chambers were identical (90/10 mmHg). In view of persistent failure to thrive he was referred to the Brompton Hospital, London, for surgical management.

Examination of the chest showed a prominent cardiac impulse at the left sternal border. The heart sounds were normal, with a grade 2/4 ejection systolic murmur in the fourth left intercostal space. The rest of the physical examination was normal.

Chest x-ray film disclosed gross cardiomegaly (cardiothoracic ratio 16/20), with prominent pulmonary vascular markings. Electrocardiogram showed sinus rhythm with a mean QRS frontal axis of +30°, left ventricular hypertrophy, and T wave inversion in leads V5 and V6. Cross-sectional echocardiography

showed situs solitus, atrioventricular concordance, and ventriculoarterial concordance. A large accessory chamber appeared to be arising from the lateral border of the left ventricle. The ejection fraction was calculated at 26%.

OPERATIVE DETAILS

Surgery was performed under deep hypothermia and circulatory arrest. The child was surface cooled to 23°C when the chest was opened via a median sternotomy incision. The left ventricle was observed to be contracting poorly. There was no external evidence of a discrete diverticulum or aneurysm. Cardiopulmonary bypass was established between a single right atrial cannula and the ascending aorta. The patient was then further cooled to a nasopharyngeal temperature of 15°C and the circulation stopped. A vertical incision was made at the apex of the left ventricle between the left anterior descending coronary artery and its third diagonal branch (Fig. 2a). The "accessory chamber" was entered and its wall appeared to contain viable myocardium. The chamber communicated freely with the main left ventricular chamber (Fig. 2b) through a large fibrous orifice (2.5 cm × 2 cm). Using a series of vertical buttressed 4/0 Prolene mattress sutures the fibrous orifice was approximated to the wall of the accessory chamber excluding the accessory chamber while preserving its functional myocardium (Fig. 2c and 2d). The ventriculotomy was then closed with two layers of continuous 4/0 Prolene (Fig. 2e). The postoperative course was uneventful. He was discharged on the tenth day taking frusemide 4 mg bd and spironolactone 8 mg bd.

Endomyocardial biopsy showed muscular hypertrophy with irregularly arranged fibroelastic tissue distinct from the regular palisades typical of endomyocardial fibroelastosis.

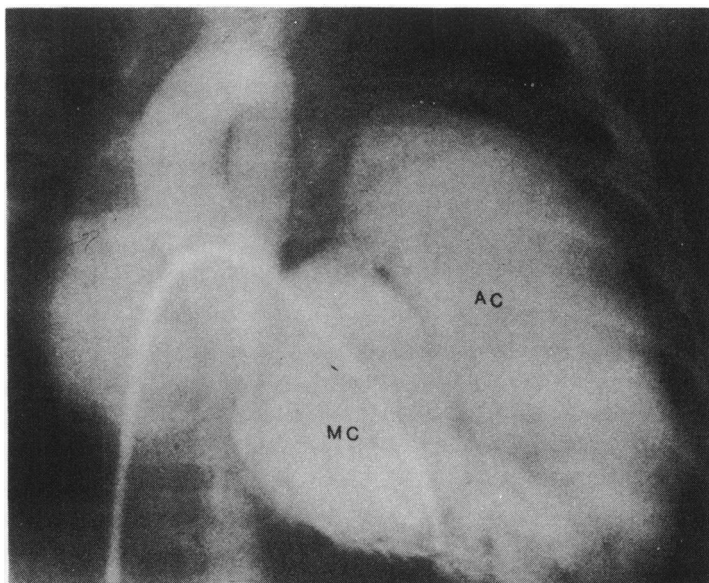


Fig. 1 Left ventriculogram showing the double-chambered left ventricle. MC, main left ventricular chamber; AC, accessory chamber.

FOLLOW UP

The child was reviewed at the age of 15 months, six months after operation. He appeared a healthy active child who was walking. His weight had increased to 11 kg (50th centile for weight). Examination of the precordium disclosed a normal cardiac impulse. The heart sounds were normal and there was no residual murmur. Chest x-ray film showed that the heart was still enlarged though the cardiothoracic ratio had reduced to 12/20. The electrocardiogram was unchanged. The left ventricular ejection fraction, calculated from repeat cross-sectional echocardiography, had risen to 44%

Discussion

Double chambered right ventricle, itself a rare condition, is created by one or more muscle bundles traversing the right ventricle.^{5,6} It is typically associated with a ventricular septal defect, and indeed the picture may be confused when the rudimentary chamber connects with the left ventricle resulting in double left ventricular chamber haemodynamics.⁷ Our case, however, was one of true double chambered left ventricle, the accessory chamber lying anterolateral to the main chamber, with the left anterior descending coronary artery and its diagonal branches running across the surface.

The aetiology of the double-chambered left ventricle is obscure, but appears to be related to a cardiomyopathy. The two early descriptions by

Paronetto and Strauss³ and Ruttenberg *et al.*⁴ reported "aneurysms" secondary to congenital cardiomyopathy. Gerlis *et al.*² suggested that endocardial fibroelastosis rendered the main chamber non-contractile and that the functional portion of the left ventricle was derived from intramyocardial sinusoids. The histology in this report was similar to the cases of cardiomyopathy reported by Alday *et al.*¹

The indication for surgery in this case was failure to thrive. The choice of operation was determined in the operating theatre, when the anatomical and pathological appearances were clearly visible. Resection of the accessory chamber was inappropriate as it would have involved excision of a large amount of viable myocardium including the left anterior descending coronary artery. Patch closure of the "ostium" between the two chambers would have resulted in too small a volume in the left ventricular cavity. Thus, a plastic procedure was chosen to maintain the orifice between the chambers widely open while obliterating the "dead space" of the accessory chamber.

There is little information on the natural history of the double chambered left ventricle. Five of the previous cases reported died, while a long term follow up on the sixth patient, treated medically, is lacking. In this particular case the operation appeared to produce a dramatic result, assessed both subjectively by the mother and objectively in the form of normal gain in weight, decrease in cardiothoracic ratio on the chest radiograph, and increase in left ventricular ejection fraction.

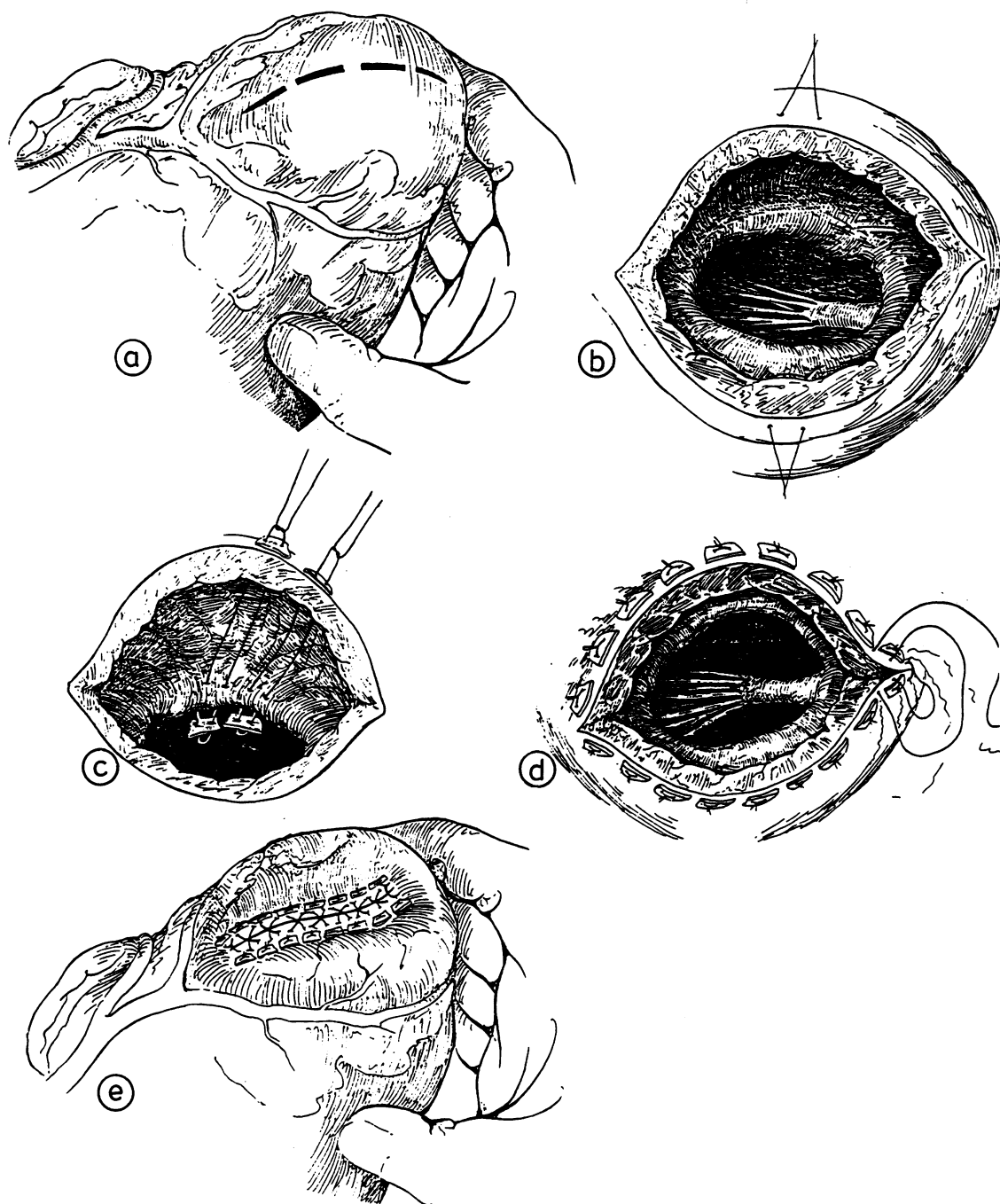


Fig. 2 (a) Vertical incision in the left ventricle between the left anterior descending coronary artery and its third diagonal branch. (b) Accessory chamber with ostium leading into true left ventricle. (c) Buttressed vertical mattress sutures inserted through the fibrous orifice and the free wall of the accessory chamber. (d) Accessory chamber excluded. (e) Left ventriculotomy closed.

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